

A fatal case of an adrenal gland melanoma with a mysterious primary lesion

A Adam, M Engelbrecht, I van Heerden

Citation

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Abstract

Adrenal gland involvement by metastatic melanoma is a common finding in patients with cutaneous or ocular melanomas. We report on the presence of an adrenal gland melanoma in a young female, without a documented primary lesion. A diagnosis of a primary adrenal melanoma was considered and after applying diagnostic criteria, could not be established. An attempt at resection via a nephroadrenalectomy and bilateral pulmonary metastectomies were performed. At 6-month follow up the patient presented with disease progression and demised prior to initiation of chemotherapy, at 22 years of age.

CASE REPORT

A 21-year-old woman presented with mild, non-specific right flank pain that was present for about four months. This was not associated with any nausea, vomiting or genito-urinary abnormalities. No other prior history of significance was noted. The general examination was unremarkable and the blood pressure was within normal limits. Abdominal examination revealed a palpable right flank mass.

Ultrasonography of the abdomen revealed an 81 x 68 mm tissue mass developed at the expense of the right adrenal gland. On further enquiry, there was no history of symptoms related to hormonal hypersecretion.

Haematological investigations including a full blood count, urea and electrolytes, inflammatory markers and liver function tests were all within normal limits. The specific hormonal levels, (including plasma catecholamines) were found to be within the physiological range. Daily urinary metanephrines and vanillylmandelic acid levels were not elevated.

The chest x-ray revealed a radiodense lesion in the lower lobe of the right lung, which was suggestive of metastasis.

On abdominal computed tomography (CT) scan, the right adrenal lesion was noted extending into the right hepatorenal recess, measuring 85 mm in its largest diameter. No evidence of hepatic metastasis, or para-aortic lymphadenopathy was detected.

Magnetic resonance imaging (MRI) of the abdomen (Figure

1) revealed a heterogeneous right adrenal mass without signal changes on chemical shift sequence.

Figure 1

Figure 1. MRI abdomen (T2-weighted), illustrating the right adrenal lesion. It measured 91mm by 73mm.



CT-guided needle core biopsies of the right adrenal mass

revealed melanin containing cells, which stained positive for both s-100 protein and human melanoma black antibody (HMB-45). The synaptophysin stain was negative. The above staining features excluded a diagnosis of a pigmented pheochromocytoma and was characteristic of a malignant melanoma.

No pigmented lesions were noted on the skin, unguis and mucosal surfaces. Ocular examination, including a slit-lamp assessment was uneventful. Rhinoscopy and oral examination was normal.

On further enquiry, the patient recalled visiting her general practitioner for a small skin lesion six years prior. This lesion was excised and a single suture was placed. Histology was not requested at that time. On meticulous inspection, a small scar below the right clavicle was noted.

To the patient, none of the above was significant enough to be mentioned.

Full body positron emission tomography (PET) CT scan (Figure 2) revealed an absence of uptake in any anatomically superficial structure or lymph node. Intense flourodeoxyglucose (FDG) uptake was only present in the right adrenal gland and in one distinct region in each lung field.

Figure 2

Figure 2. PET CT scan showed intense FDG uptake in the right adrenal gland with a central photopenic (necrotic) area and two distinct lung metastatic lesions.



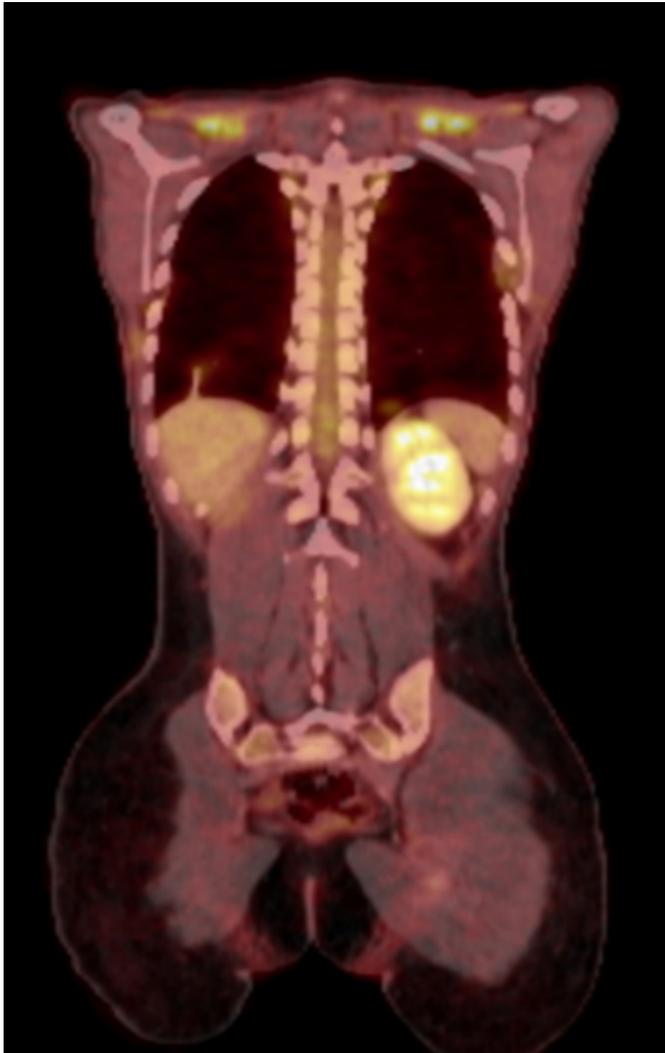
Serum lactate dehydrogenase (LDH) was elevated at 837 U/l.

An attempt at curative resection, by means of a right nephroadrenalectomy with bilateral pulmonary metastectomies was performed.

At the 6 month follow-up visit, whole body PET CT scan revealed an absence of residual lesions in the right renal bed and the lung fields bilaterally (Figure 3a).

Figure 3

Figure 3a. This coronal view illustrated the ‘macroscopic’ resection of the previous tumour burden. Post surgical changes were noted in the right lower lobe lung field.



However, other sectional views were consistent with disease progression and demonstrated widespread metastatic disease with involvement of the brain (Figure 3b), liver (Figure 3c), peritoneum, pelvic lymph nodes and both iliums.

Figure 4

Figure 3b. PET CT brain revealing the right frontal lobe lesion.

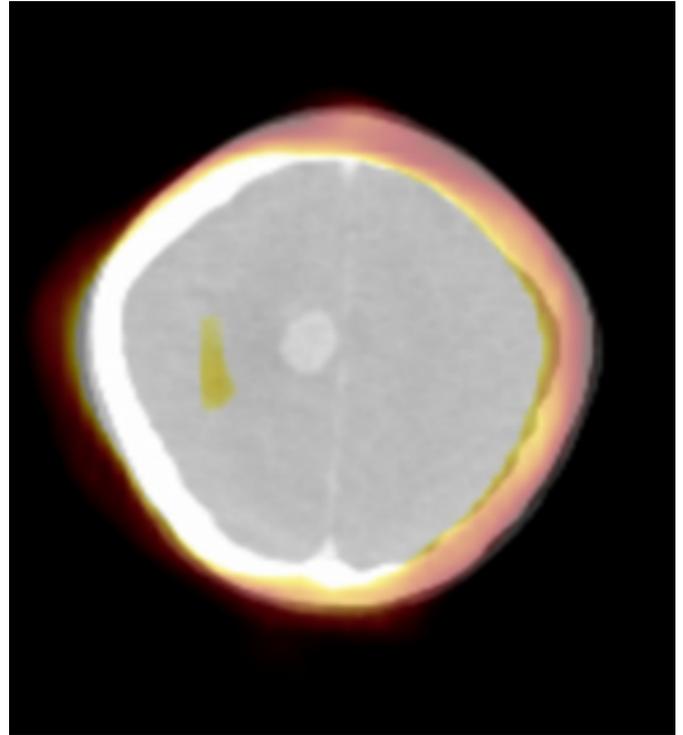
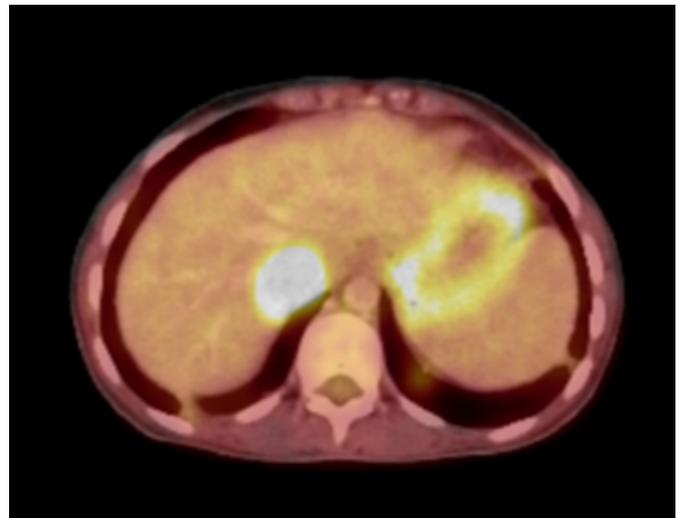


Figure 5

Figure 3c PET CT abdomen illustrating the metastatic liver lesion. It measured 3.1 x 4.0 cm.



Despite an early aggressive attempt at surgical resection, the above patient subsequently demised (7 months from initial presentation) a week prior to the initiation of chemotherapy.

DISCUSSION

Since our index patient did not have any muco-cutaneous lesions on presentation and histological assessment of the

'enigmatic' excised skin lesion was not requested, we were compelled to consider a diagnosis of a primary adrenal melanoma (PAM).

The first case of PAM was reported in a 60 year-old male in 1946.^{1,2} According to a case report discussion published in 2006, less than 20 cases of PAM were reported in the literature.¹ Since these tumours are not hormone secreting, they usually present with a locally advanced tumour load and can be aggressive.^{1,3} The longest reported tumour-free survival post initial resection was 46 months.^{1,2}

Adrenal gland involvement by metastatic melanoma can be found in up to 50 percent of patients with ocular or cutaneous melanomas. In some cases the primary lesion may have been hidden or regressed spontaneously.³ Bearing this in mind, the following rigid diagnostic criteria have been in place to diagnose PAM.¹ :

- (1) The presence of melanoma in only one adrenal gland
- (2) No history of a melanoma or pigmented lesion
- (3) Absence of endocrine disorders
- (4) Histological features characteristic of melanoma

Confirmation at autopsy is also mentioned, however this may not always be feasible.^{1,3}

PAM as a distinct possible entity, can be supported by the embryological origin of the adrenal gland.^{4,5} The adrenal medulla arises from the migration of the neural crest cells to the adrenal primordium. These neural crest cells exclusively provide the source of melanocytes to the dermis and meninges. Therefore primary adrenal lesions arising from the medulla may also produce melanin pigment, as they all migrate from the common neural crest.³

In the surgical management of isolated PAM, the procedure of choice appears to be a nephroadrenalectomy, since adhesions to the adjacent kidney is a common finding.²

Since this patient has had a lesion excised previously (and in the absence of histology of that lesion), according to the

diagnostic criteria we can not insist on a diagnosis of PAM.

In the management of metastatic melanoma, surgical resection has been the only form of treatment that has reported an actual 5-year survival and must be contemplated where appropriate.⁶ The site and number of metastatic lesions, an elevated LDH level and the disease-free-interval before metastatic development have been found to be among the most important predictors of poor outcome.⁶

CONCLUSION

The possibility of this being a metastatic lesion to the adrenal gland seems possible. Since histology was not requested on the previously excised skin lesion, we can never be certain.

PAM is a rare entity and its diagnosis can be excluded by applying certain criteria. The role of a PET study in this criteria should be contemplated in future.

Patients with metastatic melanoma may present with loin pain and a palpable flank mass.

The search for the primary lesion in metastatic adrenal melanoma may be exigent and (as illustrated in this report) it may not even be present at the time of diagnosis.

DEDICATION

This report is dedicated to the above patient, her husband and family.

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Author Information

Ahmed Adam, MBBCh Dip PEC(SA)

Registrar, Department of Urology Steve Biko Academic Hospital University of Pretoria Tswane South Africa

Matthys J Engelbrecht, FCS (Urol) (SA)

Consultant Urologist, Department of Urology Steve Biko Academic Hospital University of Pretoria Tswane South Africa

Izak J van Heerden, FCS (Urol) (SA)

Consultant Urologist, Department of Urology Steve Biko Academic Hospital University of Pretoria Tswane South Africa